School Accommodations for Sickle Cell Disease

INDIANA HEMOPHILIA & THROMBOSIS CENTER, INC.

helping patients thrive

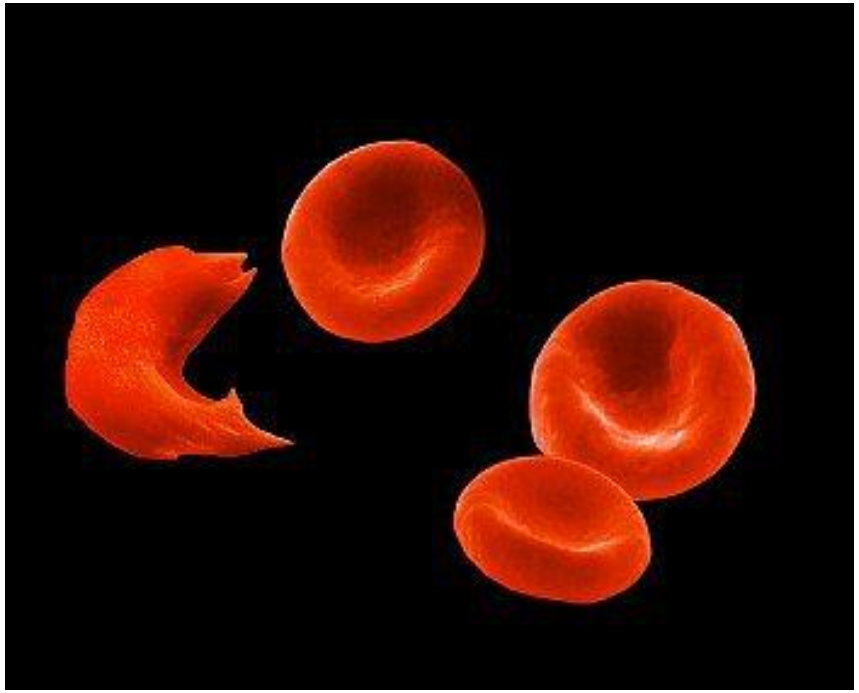
317.871.0000 or 877.256.8837

Agenda

- **What is Sickle Cell Disease?**
- **Responding to Complications at School**
- **Other Potential Health Problems**
- **Prevention: What Schools Can Do**

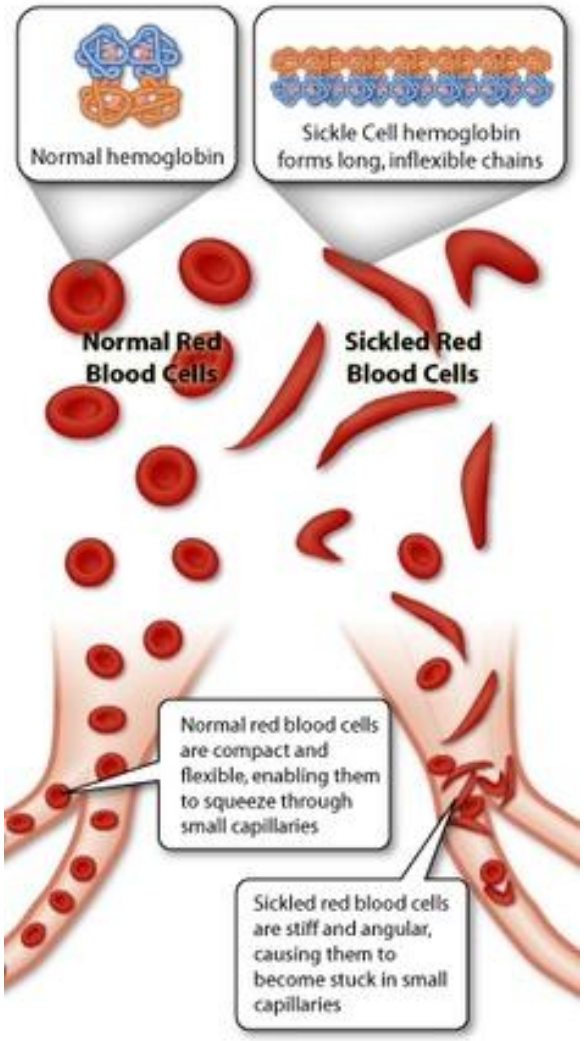
What is Sickle Cell Disease?

- **Group of hereditary blood disorders that affect hemoglobin**
- **Normal red blood cells contain Hemoglobin A**
- **Sickled red blood cells contain Hemoglobin S**



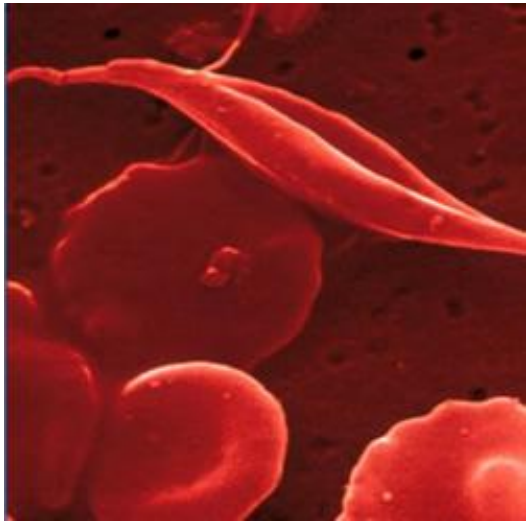
Hemoglobin S

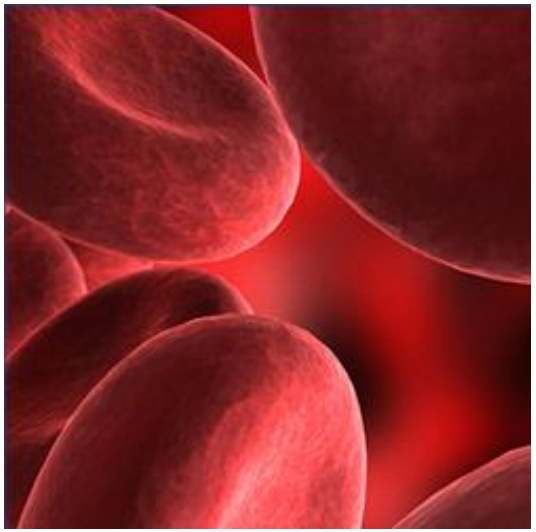
- **Polymerization of Hemoglobin S results in red blood cells that are rigid, sticky, and fragile**
- **RBCs with Hgb S have shorter life span and trouble passing through blood vessels**



Why is it Called Sickle Cell Anemia?

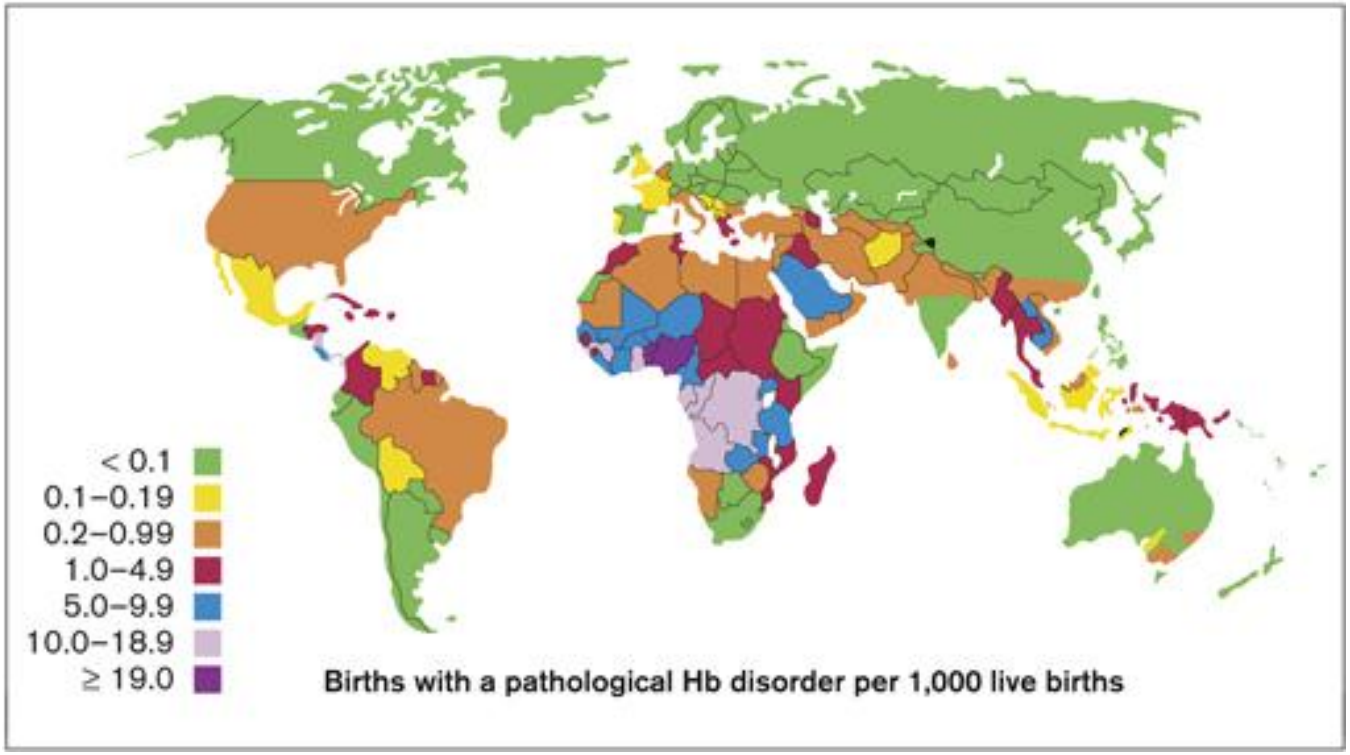
- **Because of their fragility, sickle cells burst (hemolytic anemia)**
  - This lowers the red blood cell count
- **Sickle cells live 10-20 days in the bloodstream**
  - Normal red blood cells live 120 days
- **All blood counts may drop if the bone marrow is suppressed by**
  - Infection
  - Folic acid deficiency



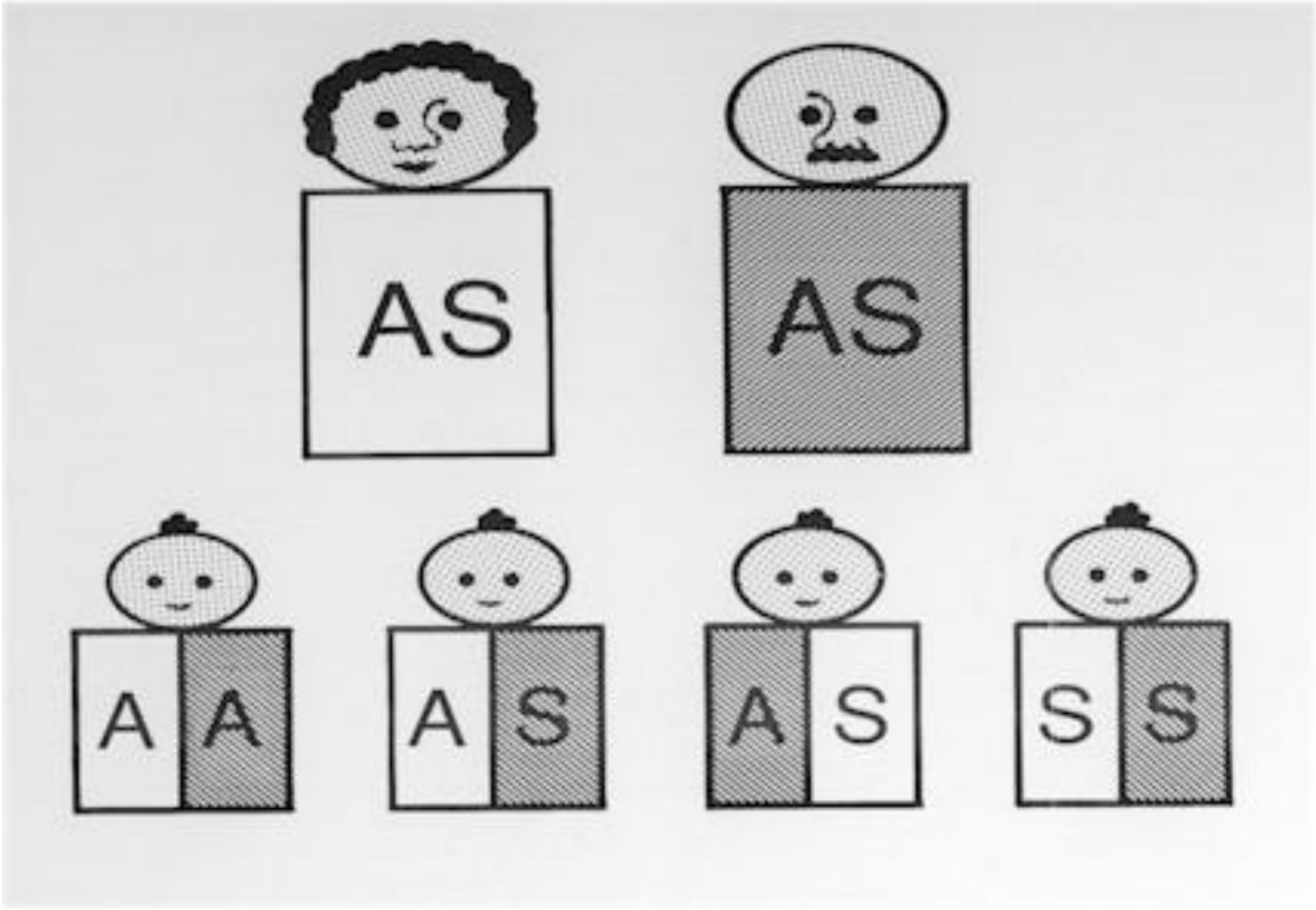


Disease Burden

- **1 in 12 African-Americans have Hgb S trait**
- **90,000-100,000 Americans have sickle cell disease**
  - ~30 children are born in Indiana each year with a hemoglobinopathy
- **Mutations arose in countries where malaria is endemic**

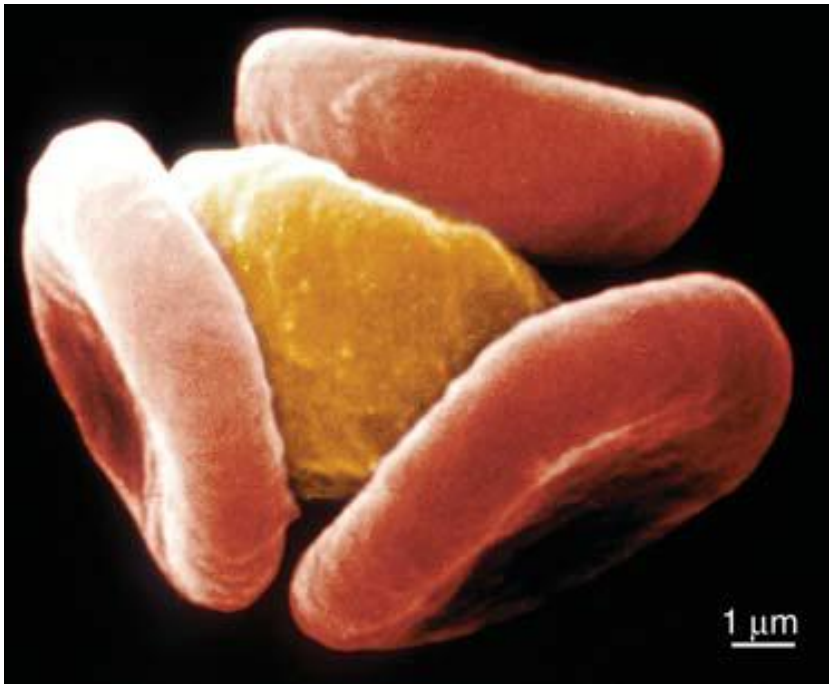


How is SCD Inherited?



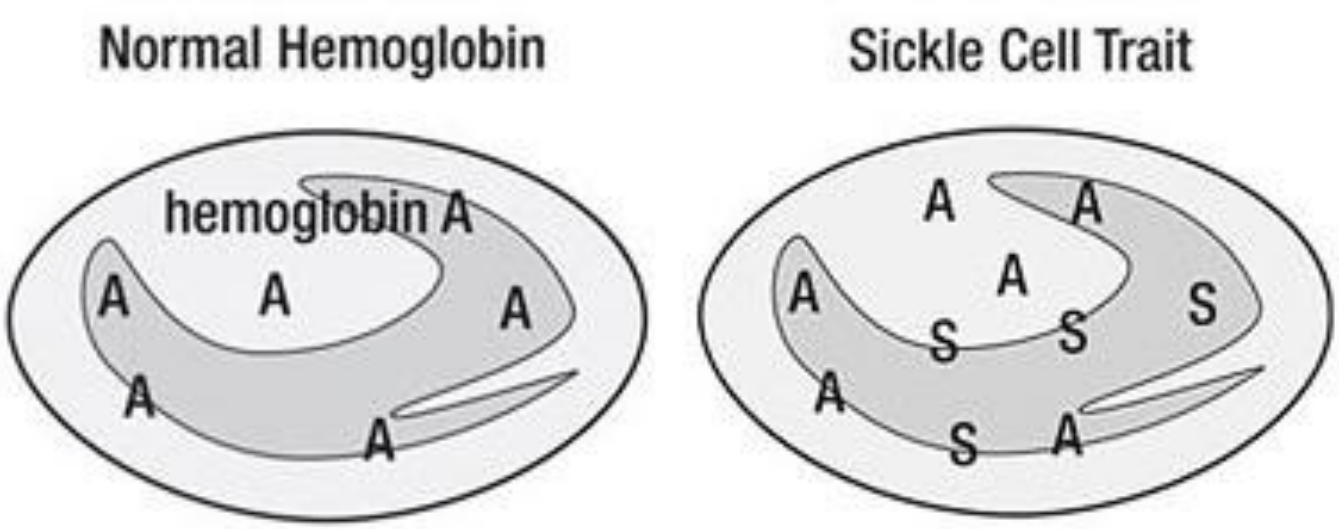
Types of Sickle Cell Disease

- **Hgb SS (Sickle Cell Anemia)**
  - ~60% of U.S. cases
  - Sub-Saharan Africa, Mediterranean, Middle East, India
- **Hgb SC**
  - ~25%
  - West and north Africa
- **Sickle β+/0 Thalassemia**
  - ~10-15%
  - Sub-Saharan Africa, Middle East, India
- **Other compound heterozygous states (Hgb SD, Hgb SE, Hgb SO)**
  - ~5%
- **Frequency and severity of complications vary by type of hemoglobinopathy**



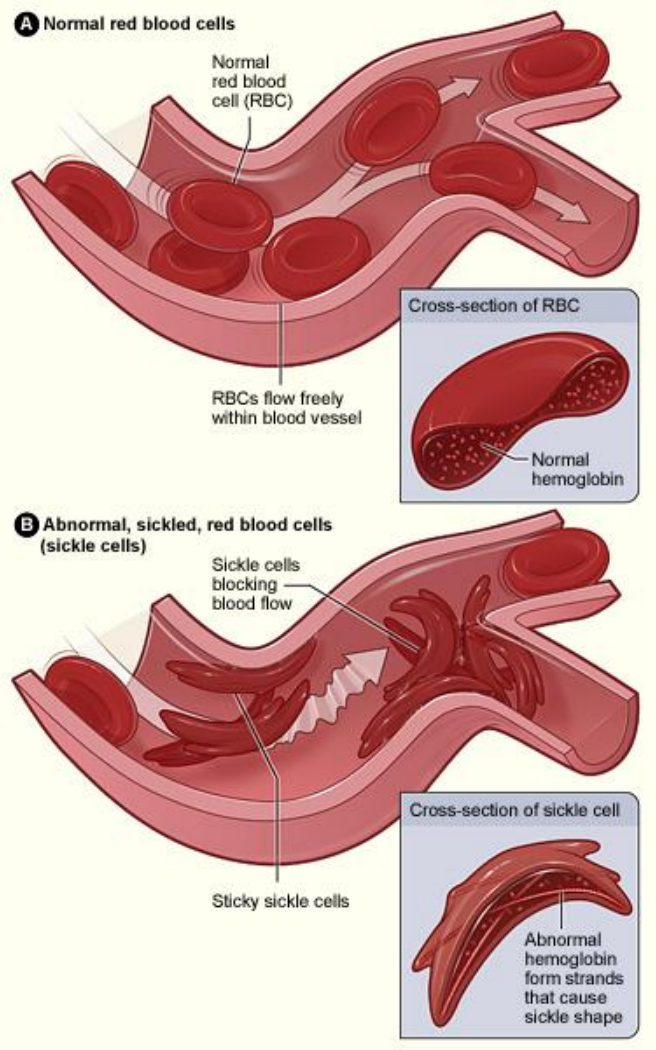
What About Sickle Cell Trait?

- **People with Sickle Cell Trait inherited Hemoglobin S from one parent and Hemoglobin A from the other**
- **Trait is a condition of the red blood cell—NOT a disease**
- **Health problems may occur under extreme conditions**
  - Dehydration, low oxygen, high altitudes



RESPONDING TO COMPLICATIONS OF SICKLE CELL DISEASE

Pain



- **Caused by sickled cells "clogging" the blood vessels**
- **Hallmark manifestation of SCD**
- **Pain can vary from mild to severe**
- **Ways to treat pain:**
  - Pain medication provided by parent
  - Heating pad
  - Hydration and rest

Pain Frequency

- **The frequency of pain episodes will vary from person to person**
- **30% of those with SCD will never or rarely have pain**
- **50% will have occasional episodes of pain**
- **20% have frequent, severe pain**
  - 6% of patients account for 30% of all pain episodes

Pain: What to Do

- **Reports of pain should be taken seriously**
  - There are usually no physical manifestations of the pain
  - Pain usually occurs in back, abdomen, arms, or legs
- **Not always necessary for child with pain to go home**
- **May only need rest, pain medication, heating pad, etc. until he or she is feeling better and able to rejoin class**
- **Distractions can also help, especially with younger children**
  - Looking at books, listening to music, toys
- **Parents should <u>always</u> be contacted when pain is first reported**

Acute Chest Syndrome

- **What Can Happen**
  - **Serious complication that can lower levels of oxygen in the blood**
  - **One of the leading causes of death in people with sickle cell disease**
  - **Symptoms include: Fever, cough, chest pain, dyspnea, hypoxia, tachypnea**

- **What to Do**
  - **Follow asthma action plan if available**
  - **Call parent immediately**
  - **If symptoms are severe, call 911**

Once a child has had one episode of ACS, they are more likely to have it again.

Asthma and Sickle Cell Disease

- **Asthma prevalence in SCD population is similar to prevalence in African-American population**
- **Asthma is associated with an increase in SCD-related morbidity and premature mortality**
- **Studies show increased incidence in painful episodes and ACS for children with asthma**
- **Children with asthma:**
  - More likely to have ACS
  - More likely to have multiple episodes of ACS
  - More likely to have ACS at a younger age
  - More likely to be hospitalized for longer with ACS

Asthma and Sickle Cell Disease

- **Asthma triggers are often sickle cell pain triggers**
  - Cold weather
  - Exertion
  - Strong emotions

Optimizing asthma control can help to control symptoms of sickle cell disease

Headache

- **What Can Happen**

- **Sickled cells tend to "clump up" along the walls of the large arteries going to the brain**
  - Damages vessel walls
  - Exposes tissue that collects more sickled cells and narrows the vessels even further
- **5-10% of children with Hgb SS will have an overt stroke**

- **What to Do**

- **F.A.S.T.**
  - Face: Any facial weakness or drooping?
  - Arm: Can the student lift both arms above their head?
  - Speech: Can the student speak clearly and understand what you say?
  - Time: To call 911 if any of these are present
- **Call parent immediately**

Priapism

- **What Can Happen**
  - Sustained, painful, unwanted erection
  - Failure of venous outflow due to sickling of blood cells
  - ~30% of males with SCD under age 20 have had at least one episode of priapism
  - Can cause impotence
- **What to Do**
  - Give pain medication provided by parent
  - Heating pad
  - Push fluids
  - Call parent if erection does not go down within 30 minutes

Infection

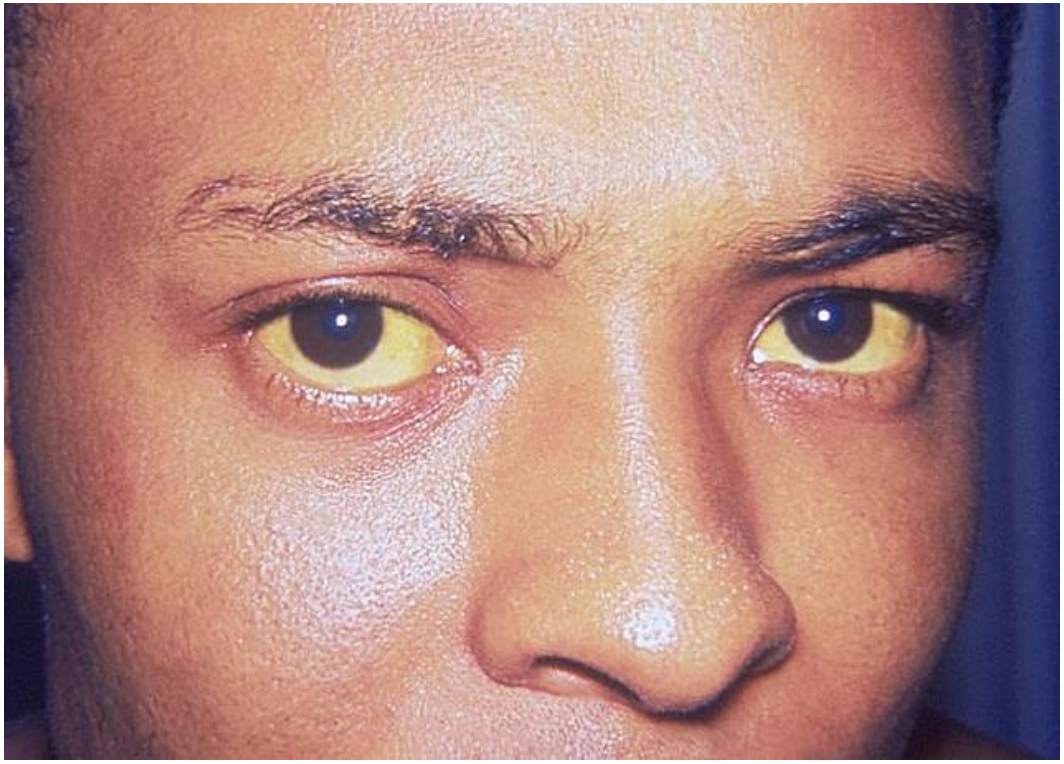
- **What Can Happen**
  - **SCD impairs splenic function, resulting in eventual functional asplenia**
  - **Without the spleen's filtering function, local infections can readily become systemic**
- **What to Do**
  - **Any fever of 101° or higher is a medical emergency**
  - **Do not give fever reducers**
  - **Call parent immediately**

OTHER POTENTIAL HEALTH PROBLEMS

Other Potential Health Problems

Gallstones and Jaundice

- **Gallstones occur in about a third of children with SCD**
- **Students with SCD may be self-conscious about jaundiced skin or eyes**



Other Potential Health Problems

Delayed Growth and Puberty

- Puberty is often delayed in children with SCD
- Children and teens with SCD may be small and thin for their age
- Usually reach full height by age 20

Retinopathy

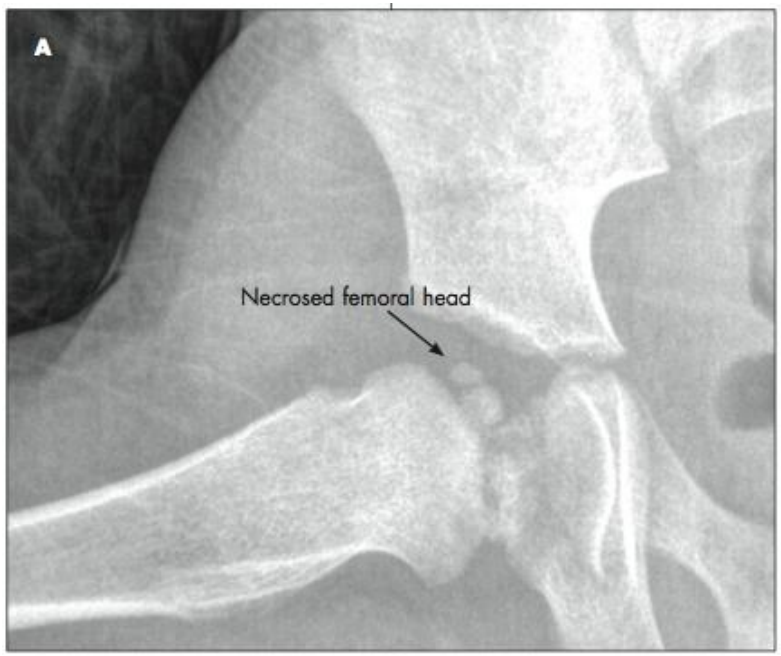
Sleepiness

- Anemia/Sleep apnea

Other Potential Health Problems

Avascular Necrosis

- Caused by reduced blood flow to the joints
- Usually affects hip or shoulder joints
- Can cause severe pain
- May affect student's ability to participate in gym activities or walk to and from classes



PREVENTION: WHAT SCHOOLS CAN DO

Plan for Extreme Temperatures

- **Alternative indoor recess when 40° F or below**
- **Access to coat or jacket during fire drills**
  - Students may need to return to the school sooner than other students, or have access to climate-controlled vehicle
- **Access to sweater or extra layers when in the classroom**
- **Offer at least one cup of water per hour when outside in the heat**
- **Bus schedules or routes may need to be changed to ensure that children with SCD do not have to walk long distances or wait a long time for the bus in cold**

Avoid Dehydration

- **Unlimited access to water or other fluids**
  - Some children may need to be encouraged to drink
- **Unlimited restroom breaks**
  - Needed due to increased fluids as well as damage to the kidneys from the sickled cells

Prevent Fatigue and Pain

- **Allow frequent breaks during gym activities and sports events, as requested by the student**
- **Listen to and act quickly upon reports of pain or headaches**
- **Never apply ice to cuts or bruises**
- **Use caution with water activities/swimming**
  - Consult with patient's hematologist for guidelines
- **Provide two sets of textbooks**
- **Sickle Cell Trait**
  - Strenuous activities

Special Accommodations

- **Studies have shown that children with SCD miss an average of 20-40 school days a year**
  - Participants in one study missed an average of 12% of the school year, with 35% of students missing at least one month of school
  - Absences may be due to pain crises treated at home, serious health complications requiring hospitalization, or frequent medical appointments

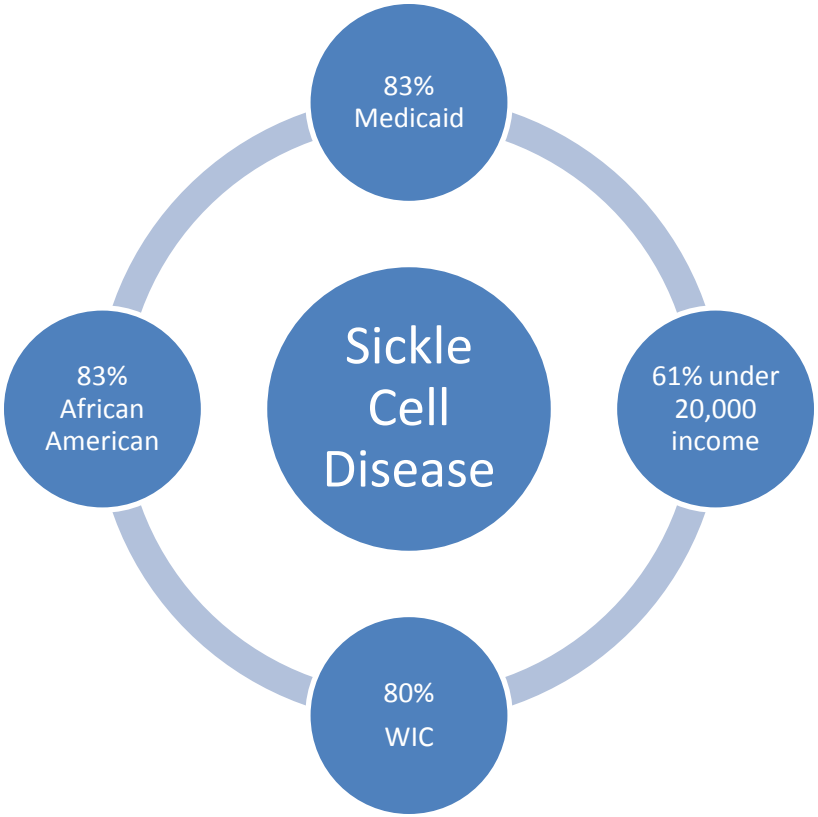
Special Accommodations

- **Students may require 504/IEP**
  - May include accommodations such as providing two sets of textbooks (one at home, one at school), plans for getting homework assignments to the child, testing/ISTEP accommodations
  - Educational services in the home/hospital
- **Due to the unpredictable nature of SCD and the varied disease course, accommodations should be tailored for each child, but every child should have an individualized health care plan**

SCD and Learning Difficulties

- **Children with SCD may struggle in school due to damage to brain tissue**
  - Overt strokes
  - Silent strokes (up to 30% of children with SCD)
  - Hypoxia from anemia, poor pulmonary functioning, sleep apnea
- **Report unexplained sleepiness or trouble concentrating to parents**
- **May need a referral for neurodevelopmental testing**

Living with Sickle Cell Disease



Sickle SAFE Program, March 2014

Living with Sickle Cell Disease

- **School personnel must be aware of the stigma surrounding sickle cell disease**
  - Students may be ashamed of diagnosis
  - Students may feel self-conscious about physical manifestations of SCD, such as jaundice or delayed growth and puberty
  - Students may worry about being different than their peers, or being a burden to their families
- **People who experience frequent painful crises are more likely to have:**
  - Low self-esteem
  - Anxiety and depression
  - Poor school performance
  - Isolation
  - Decreased participation in activities of daily living
  - Poor peer and family relationships

The Child with Sickle Cell Disease. . .

- **Should be taken seriously when they present with any reports of pain or fatigue**
- **Should be encouraged to participate in school activities as much as possible**
- **Should be discouraged from thinking of themselves as “sick” or less capable than other children**

Handout Requests

- **Please email me at ebloom@ihtc.org to request electronic copies of today's handouts**

Contact Information

Ellen Bloom, MPH, CHES

Newborn Screening Program Coordinator

Indiana Hemophilia and Thrombosis Center

8402 Harcourt Road Ste. 500

Indianapolis, IN 46260

317-871-0011 ext. 292

ebloom@ihtc.org